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Hylonome

Clinical Quiz

Persistent knee effusion in a young female

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Case

A 14-year-old female with painless joint effusion in the right knee was referred to our rheumatology department from the orthopaedics department. The patient first noted right joint swelling 20 days prior to referral with no history of trauma or a change in limb function. Prior to referral, the patient received non-steroidal anti-inflammatory drugs after which the joint was aspirated for the purposes of drainage. No analysis of the aspirate was performed.

On clinical examination, the right knee joint was warm, full knee flexion was limited, and palpation revealed a supra patellar effusion. The joint was not red and no skin changes were noted. No other systemic signs were observed. Joint aspiration revealed clear synovial fluid. Routine methylprednisolone was injected into the joint immediately after aspiration. On cytology of the synovial fluid, cell count and type were within normal limits and cultures were negative. All immunological, haematological and biochemical tests were within normal limits.

An U/S was done two weeks later due to recurrence of symptoms and synovitis of the joint was noted. A T2/sequence MRI revealed scattered areas of low signal intensity resembling haemosiderin deposition and hyperplastic thickening of the synovium.

Arthroscopy of joint synovium revealed a mixture of rusty brown and white hyperplastic villi. The biopsy showed infiltration of multinucleated giant cells confirming the diagnosis of Pigmented Villonodular Synovitis. Three months later, a similar picture was seen on the left knee. MRI, arthroscopy and synovial biopsy showed similar findings on the left knee.

The lesions were treated by surgical resection with total synovectomy followed by radiosynoviorthesis with yttrium 90 to cover for any unresected tissue.

Commentary

Pigmented villonodular synovitis (PVNS) is a rare benign proliferative disorder that affects the synovium of joints[1]. The nomenclature and description of PVNS has changed over the years. Chassaignac was the first to record PVNS in 1852, and described it as a nodular lesion of the synovium, which affects the flexor tendons of the phalanges. Jaffe and colleagues made the current formal description in 1941. PVNS has an indeterminate multifactorial aetiology. Neoplastic changes, genetic anomalies involving rearrangement of genes and an inflammatory response to the synovium are all currently accepted theories[2].

The symptoms that suggest PVNS vary and depend on whether the lesion occurs locally or diffusely. It most frequently presents as a painful swelling, or soft tissue mass of acute onset, which is out of proportion to the degree of discomfort. More than a third of patients have a history of trauma[2]. The diffuse form, which is the most frequently seen, affects the bursa, tendon sheath and synovium of large joints with a predilection for the knee joint in 80 % of cases[2,3]. The prevalence is about 1.8 cases of intra-articular disease and 9.2 cases of extra-articular disease per 1 million, respectively[4]. Multifocal manifestation is rarely seen but seems to appear more frequently in children[3,4]. Patients most commonly affected are in their 3rd to 5th decade of life. Rarely, children are affected and they present with a range of other abnormalities such as Noonan syndrome, extreme lymphedema, cherubism and vascular lesions. Many studies have shown males and females to have an equal prevalence[1,4].

On gross pathology there is hypertrophy of the synovium with thickening of the joint usually accompanied with nodular protrusions and villous projections. The thickened joint appears spotty yellow and dark brown due to xanthoma cells and haemosiderin deposits. The synovial fluid usually has a bloody to yellow appearance[1,4].

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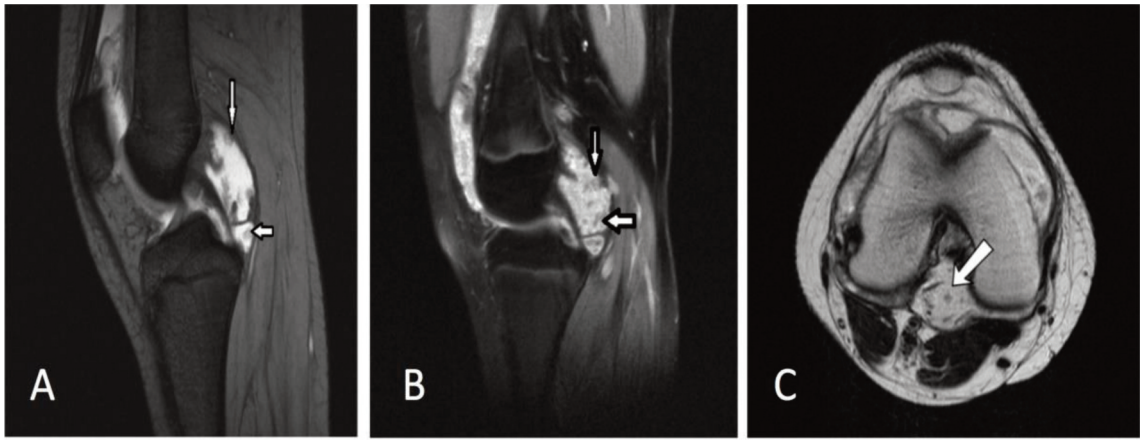

Figure 1. A. Arrows showing a mixed signal pattern on a T2 weighted MR imaging of the right knee with corresponding areas of haemosiderin. **B.** T2 weighted MRI of the left knee showing abnormal high and low signals within tissue. These areas correspond to haemosiderin-laden synovium. C. Axial magnetic resonance imaging (MRI) scan at the level of the knee joint shows mixed signal pattern.

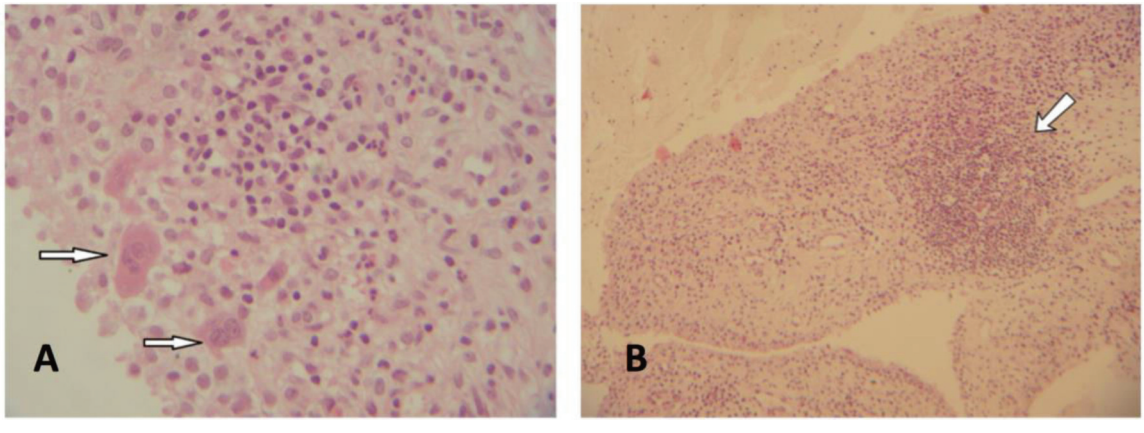

Figure 2. A. Histology of the right knee showing multinucleated giant cells. **B.** Synovium of the right knee showing infiltration of inflammatory cells.

Histologically there is infiltration of the synovium by mononuclear stromal cells, multinucleated giant cells and xanthoma cells.

Radiographic images are non-specific. They may show soft tissue swelling, and bone erosion with the maintenance of the joint space. CT normally shows synovial thickening with bone erosions. MRI shows the severity of the disease. Blooming of low signal intensity is pathognomonic for PVNS. It is seen because of artifact created when the magnetic field detects the haemosiderin deposits, frequently found in the lesions. Sonography helps with identifying any extra articular lesions of the surrounding tissues. A correct diagnosis of PVNS is usually made with a correlation of the imaging findings and the histological features[4].

The isolated finding of a non-traumatic joint, should lead to the consideration of an alternative diagnosis. Differential diagnoses include rheumatoid arthritis, systemic lupus erythematosus, psoriatic arthritis, osteoarthritis or pigmented villonodular synovitis[2].

Surgical resection with total synovectomy is the treatment of choice. In cases of recurrence adjuvant radiotherapy is a treatment option. Incomplete surgical resection and disease in joints are directly related to higher recurrence rates if untreated. PVNS results in pain, loss of function and destruction of the affected joint. Recurrence has been shown to cause arthritis mutilans[4].

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Questions

1. Select from the options below, the one most helpful in establishing a diagnosis of PVNS

A. MR imaging findings of predominant low signal intensity with hemosiderin content.
B. A combination of plain radiographs, MRI findings and histology from biopsied tissue.
C. A combination of plain films and MRI only
D. A combination of plain films, MRI findings and CT scan findings
E. Bone scans

Critique

Histological features of atypical cells and the presence of multinucleated giant cells in conjunction with the imaging findings helps to confirm the diagnosis.
The correct answer is B.

2. Which of the following joints is most frequently affected by pigmented villonodular synovitis:

A. Metatarsophalangeal joint.
B. Temporomandibular joint.
C. Apophyseal joints of the spine.
D. Knee.
E. Elbow.

Critique

PVNS can occur diffusely or locally. The diffuse form of PVNS is the most frequently seen and affects the large joints, with the knee involved in 66-80% of cases. Rarely affected joints include the sacroiliac joint, subtalar joints, temporomandibular joints, small joints of the hands and feet and the apophyseal joints of the spine. The elbow is the least affected of the large joints.
The correct answer is D.

3. Which of the following best describes the initial treatment option for PVNS?

A. Surgical resection with complete synovectomy.
B. Corticosteroids.
C. Corticosteroids and radiosynovectomy.
D. Radiosnyovectomy.
E. Surgical synovectomy and radiosynovectomy.

Critique

Surgical resection with complete synovectomy is the treatment of choice in all types of PVNS. It has been observed that recurrence occurs frequently in diffuse intra-articular PVNS, in which case an adjuvant radiation therapy may be added to the treatment regime.
The correct answer is A.